



Chiari's Network: Association with Other Congenital Heart Diseases

Chiari Ağı: Diğer Konjenital Kalp Hastalıkları ile İlişkisi

Chiari Network in Children

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Özet

Amaç: Bu çalışma ile pediyatrik kardiyoloji polikliniğine müracaat eden çocuklardaki Chiari Ağı'nın (CA) transtorasik ekokardiyografi ile prevalansını değerlendirmesi ve diğer kardiyak anomalilerle birlikteliğinin belirlenmesi amaçlanmıştır. **Gereç ve Yöntem:** Hastanemizin çocuk kardiyoloji bölümünde Nisan 2013-Nisan 2014 yılları arasında kardiyak muayene ve transtorasik ekokardiyografi uygulanan 2232 çocuk çalışmaya alındı. Rutin M-mode, 2-D ve Doppler uygulandı. Tüm eşlik eden lezyonlar kaydedildi. Erken ve geç diastolik akım hızı zirve yüksekliklerinin (E/A) 1'in altında olması muhtemel sağ ve sol ventriküler diastolik disfonksiyonu düşündürdü. **Bulgular:** 2232 çocuktan 76'sında CA tespit edildi (%3.4) ve en sık ilk bir ayda saptandı. CA ile birlikte en sık görülen anomaliler atrial septal defect (ASD) ve patent foramen ovale (PFO) idi (%55.3). 10 hastada (%13.2) periferik pulmoner stenoz (PPS) ve 7 hastada (%9.21) atriyal septal anevrizma (ASA) tespit edildi. 2232 hastanın 267'sinde (%8.4) ASA saptanırken CA'lı hastalarda ASA sadece 7 kişide (%9.6) saptandı. **Tartışma:** CA sık görülmeyen, tesadüfi olarak tespit edilebilen ve doğru bir şekilde tanısı konması gereken, her zaman masum olmayan bir defektir. Bu her zaman normal bir varyant olmayabilir. İlerde olabilecek muhtemel komplikasyonlar nedeniyle Chiari ağı uygun bir şekilde dökümente edilmelidir.

Anahtar Kelimeler

Chiari Ağı; Çocuk; Konjenital Kalp Hastalığı

Abstract

Aim: The study was performed to assess the prevalence of Chiari's Network (CN) in a pediatric outpatient department population by using transthoracic echocardiography (TTE) and to determine its association with other cardiac anomalies. **Material and Method:** 2232 children who underwent cardiac examination and TTE at the pediatric cardiology outpatient department of our institution between April 2013 and April 2014 were included in the study. Routine M-mode, 2-D, and Doppler studies were applied. All co-existent lesions were recorded. Ratio between heights of early and late diastolic flow velocity peaks (E/A) were <1 in tricuspid and mitral valves considered as possible right and left diastolic dysfunctions respectively. (Bu cümle eklendi.) **Results:** Of 2232 children, CN was detected in 76 patients (3.41%) and found most frequently during the first month. Atrial septal defect and patent foramen ovale (ASD/PFO) was the most common congenital defect seen with CN (55.3%). Ten children (13.2%) had peripheral pulmonary stenosis (PPS) and seven (9.21%) showed atrial septal aneurism (ASA) associated with CN. Of patients with CN, a total of 11 patients (14.5%), 10 of them 1-8 days old and 1 of them 17 days old, had E/A<1 in tricuspid valve. Three (3.9%) patients 1-12 days old demonstrated E/A<1 in tricuspid and mitral valves. Twenty-nine of the studied patients (11%) demonstrated E/A<1 in tricuspid valve (90% 1-30 days old). Of the total 2232 patients, 267 individuals (8.4%) showed ASA whereas there were only 7 cases (9.6%) of CN patients. **Discussion:** CN is an uncommon and incidental finding that should be recognized appropriately. This may not be always a normal variant. Proper documentation of CN is critical regarding possible future complications.

Keywords

Chiari's Network; Children; Congenital Cardiac Anomalies

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Introduction

Chiari's Network (CN) is a fairly rare defect that was first described by Hans Chiari [1] in 1897. In fetal life, the Eustachian valve, which normally regresses at 9–15 weeks of gestation, is located in the superior portion of the right sinus venosus and directs blood from the inferior vena cava toward the foramen ovale. Incomplete regression causes a persistent or prominent Eustachian valve. It can be determined by the presence of a thin ridge that arises from the anterior rim of the inferior vena cava orifice [2]. If the Eustachian valve remnants involve a membrane of fine fenestrated fibers that is located in the right atrium, it is called CN [1,3,4,5].

CN is usually an incidental finding with no further clinical consequences. Usually it has no clinical significance. Nevertheless, a number of cases have been described with clinical pathology believed to be associated with the presence of CN [6,7,8,9]. It has been reported that it can cause thromboembolism, infective endocarditis, and dysrhythmias, and that it represents a physical barrier to invasive procedures [10]. In light of this information, we aimed to study the prevalence of CN and whether it is associated with other congenital defects among our pediatric cardiology outpatient department records.

Material and Method

All patients who underwent cardiac examination and trans-thoracic echocardiography (TTE) at the pediatric cardiology outpatient department of our institution between April 2013 and April 2014 were included in the study. Routine M-mode, 2-D, and Doppler studies were applied. All co-existent lesions were recorded. Ratio between heights of early and late diastolic flow velocity peaks (E/A) were <1 in tricuspid and mitral valves considered as right and left ventricular diastolic dysfunctions respectively. CN was diagnosed by TTE, defined as a remnant originating close to the Eustachian or Thebesian valve at the orifice of the inferior vena cava or the coronary sinus and demonstrating attachments to the upper wall of the right atrium or the interatrial septum (Fig 1, Fig 2, Fig 3). All patients were studied regarding the coexistence of other congenital anomalies with CN.

Results

Of 2232 studied patients, CN was detected in 76 patients (3.41%). The male/female ratio among patients with CN was 43/34. The co-lesions associated with CN and age distribution of these patients are listed in Tables 1 and 2 respectively. Atrial septal defect and patent foramen ovale (ASD/PFO) was the most common congenital defect seen with CN (55.3%). It was found most frequently during the first month. Of the total studied population, 267 patients (12%) demonstrated atrial septal aneurysm (ASA) whereas 7 patients (9.6%) had CN. The characteristics of the CN patients with peripheral pulmonary stenosis (PPS) are listed in Table 3. The age distribution of all ASA patients is shown in Table 4. The co-lesions with ASA are summarized in Table 5. PFO was the most common defect observed with ASA. Of patients with CN, a total of 11 patients (14.5%) (10 of them 1–8 days old and one patient 17 days old), and 3 patients (3.9%) all 1–12 days old, demonstrated a ratio between heights of early and late diastolic flow velocity peaks



Figure 1. Chiari's Network originating beneath coronary sinus toward tricuspid valve (White arrow)



Figure 2. Chiari's Network originating above the coronary sinus passing tricuspid valve and floating in right ventricle (orange arrows)

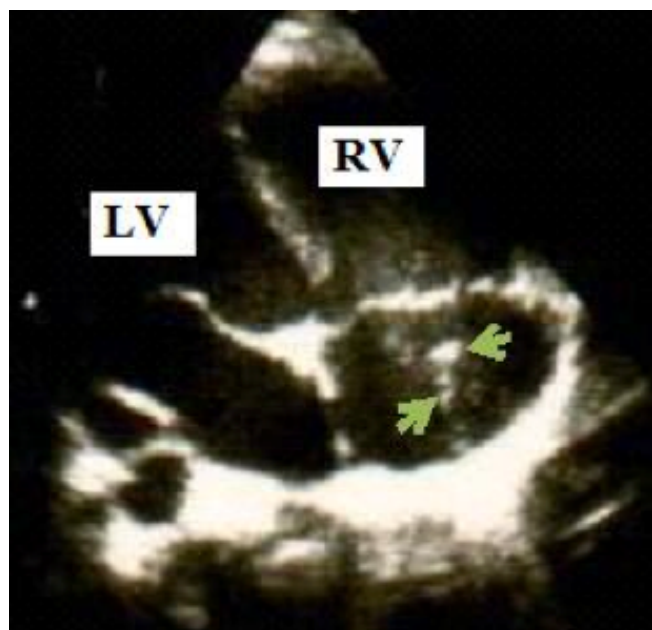


Figure 3. Chiari's Network originating superior vena cava orifice floating in right atrium (green arrows)

(E/A) less than 1 over the tricuspid and mitral valves, respectively. This was true for only the tricuspid valve of 29 out of 267 patients with ASA (11%) (90% 1-30 days old). The latter findings indicate that further studies are necessary to rule out

Table 1. Co-Lesions associated with Chiari's Network (n=76)

Co-Lesion	Number	%
ASD / PFO	42	55.3
PPS	10	13.2
ASA	7	9.21
LV band	7	9.21
VSD	5	6.58
PS	2	2.63
MVP	2	2.63
MVI	2	2.63
LSVC	1	1.32
PVR	1	1.32
ToF	1	1.32
No co-lesion	16	21.1

ASA: atrial septal aneurism, ASD: atrial septal defect, PFO: patent foramen ovale, CoAo: coarctation of the aorta, IVSH: intraventricular septal hypertrophy, LSVC: left superior vena cava, LV: left ventricular, MVP: mitral valve prolapsus, MVI: mitral valve insufficiency, PDA: patent ductus arteriosus, PPS: peripheral pulmonary stenosis, PS: pulmonary valve stenosis, PVR: pulmonary vascular resistance, ToF: tetralogy of Fallot, VSD: ventricular septal defect

Table 2. The age of patients with Chiari's Network (n=76)

1-29 days	29
1-12 months	24
13 - 60 months	12
61 months - 10 year	9
> 10 year	2

Table 3. Age and gender distribution of Chiari's Network patients with peripheral pulmonary stenosis and co-lesions (n=10)

Sex	n
Male	5
Female	5
Age	
1-29 days	6
30 - 60 days	2
> 60 days	2
Co-lesions	
ASD/PFO	7
ASA	2
PDA	1
VSD	1
MVI	1

ASA: atrial septal aneurism, ASD: atrial septal defect, PFO: patent foramen ovale, PDA: patent ductus arteriosus, MVI: mitral valve insufficiency, VSD: ventricular septal defect,

Table 4. Age distribution of patients with atrial septal aneurysm (n=267)

Age	n	%
1-30 day	143	54
1-12 months	88	33
1-5 years	24	9
> 5 years	12	4

Table 5. Co-lesions observed in patients with atrial septal aneurysm (n=267)

Co-lesion	n	%
PFO	95	36
ASD	79	3
PPS	50	19
CN	49	18
VSD	33	12
PDA	32	12
PS	19	7
LV fibrous band	19	7
MY	4	1
CoAo	2	1
LSVC	2	1
Bicuspid aorta	2	1
Septal Hypertrophy	1	
ToF	1	

ASD: atrial septal defect, PFO: patent foramen ovale, CoAo: coarctation of the aorta, LSVC: left superior vena cava, PDA: patent ductus arteriosus, PPS: peripheral pulmonary stenosis, PS: pulmonary valve stenosis, ToF: tetralogy of Fallot, VSD: ventricular septal defect, LV: left ventricular, MY: mitral regurgitation.

the possible coexistence of any ventricular diastolic dysfunction among pediatric patients, especially the neonates with CN and ASA.

Discussion

Developmentally, the embryological remnants of the right venous valve evolve into the valve of the inferior vena cava (Eustachian valve) and the valve of the coronary sinus (Thebesian valve) and Chiari Network (CN) [11,12].

Echocardiography is an excellent tool for the diagnosis of these structures, including CN. In transthoracic and transesophageal echocardiography, CN presents as a highly mobile, highly reflective echo target in several locations in the right atrium [13]. CN is believed to be of little clinical consequence. However, in previous reports it has been shown that it is not always a benign structure [14]. This structure, particularly in the context of fever, congestive heart failure, pulmonary infiltrates, or a history of intravenous drug abuse, could be mistaken for evidence of active infection or disruption of normal right-sided structures, possibly requiring urgent cardiac surgery [13]. It may cause persistence of a patent foramen ovale and formation of an atrial septal aneurysm and may facilitate paradoxical embolism [15]. It may be associated with thrombi formation, and part of the strands may be embolized [16,17]. Infective endocarditis has been reported in association with the CN [18,19]. It also may cause abnormal atrial depolarization favoring supraventricular dysrhythmia [20]. Catheters [21], guidewires [22], and pacemaker leads [23] may become entrapped within the network during invasive procedures. CN has also been demonstrated in association with neurofibromatosis [24], Behçet syndrome [25], platypnea-orthodeoxia [26], and migraine [27] and therefore it is not always a benign structure [28].

In the current study, the prevalence of CN in the pediatric cardiology outpatient department of our hospital was 3.4%, which is compatible with other published reports. Previous studies have reported the prevalence to be approximately 2% [29]; 4.6% as a postmortem finding [30]; and 2% as screened by TTE. These

studies report no significant difference regarding age and gender [15].

CN may be associated with an increased prevalence of other congenital anomalies including a PFO and ASA [31, 15]. The etiology is not well known, but coexistence with other congenital heart disease is well documented [2,3]. Schneider et al. [15] demonstrated the presence of a PFO in 83% of patients with CN as compared to 28% in control patients among 1436 adult patients evaluated by TEE. The current study showed that ASD and PFO were the most common (55.3%) accompanying findings among individuals with CN. Additionally CN was found frequently among patients with ASA. It has been reported that ASA was found in 24% of patients with CN diagnosed by TEE [15]. We observed ASA in about 9.2% of our patients and it occurred most frequently during the first month of life. Ninety-five percent of our patients with ASA were associated with PFO. In a study evaluating patients with embolic stroke, of those with a PFO and ASA, a significant number (6/32) showed coexistence with CN [32]. ASA has been attributed to a congenital connective tissue defect, leading over time to degeneration and weakening of the interatrial septum with subsequent aneurysm formation.

PPS in our series was also associated with CN (13.2%), mostly (80%) among patients less than 2 months old. It may produce continuous or systolic murmur, clinically, that could be confused with bruit de Roger of a ventricular septal defect [32] and characterized by multiple distributed narrowed ostia of the branch pulmonary arteries. It is frequently associated with Alagille and Williams syndromes and can result in pulmonary flow disparity and right ventricular hypertension [33]. In clinical practice, various symptoms can be observed depending on the severity of stenosis and coexisting cardiac and pulmonary disorders. Some cases may mimic pulmonary embolism, leading to misdiagnosis. We detected tricuspid $E/A < 1$ in 11 (14.5%) and also $E/A < 1$ for mitral and tricuspid valves in 3 (3.9%) of our patients with CN, whereas 29 individuals out of 267 (11%) with ASA showed $E/A < 1$ in tricuspid valve. We were unable to find any information about coexistence of CN or ASA with $E/A < 1$ in tricuspid valve in the pediatric age group.

Conclusion

CN, not always an innocent abnormality, is an uncommon and incidental finding that should be recognized appropriately. Proper documentation of CN is critical regarding possible future complications.

Competing interests

The authors declare that they have no competing interests.

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